

Left atrial myxoma as a cause of multiple cerebral microembolization (RCD code: VI-1A.1)

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Abstract

We report a case of a 56-year-old female after breast cancer treatment, who was diagnosed with left atrial myxoma as a rare cause of cerebral and cerebellar stroke. JRC D 2017; 3 (5): 168–170

Key words: cardiac tumor, cerebral stroke, rare disease

Case presentation

We present a 56-year-old female with hypertension, hyperlipidemia, after mastectomy, chemotherapy and radiotherapy due to breast cancer who was admitted to the Neurology Department in October 2016 with a suspicion of cerebral stroke after a sudden fall without loss of consciousness.

Neurological examination revealed that the patient was alert, attentive, oriented and cooperative with normal speech but responses were slow and the patient was somnolent. Meningeal signs were negative. She presented with exotropia of the right eye, pupils were bilaterally equal in size with normal reaction to light. Reduced motion speed of left limbs, slightly decreased muscle force and positive Romberg's test were observed. Physiological reflexes were normal, with negative Babinski sign bilaterally.

Laboratory tests revealed slightly elevated troponin T of 0,038 (N: <0,014 ng/ml), white blood cell count of 10.4 (N: 4.3–10.0 × 10⁹ /l), neutrophils of 9,0 (N: 2.5–5.0 × 10⁹ /L), C-reactive protein of 34 (N: <5 mg/l), d-dimer: 560 (N <500 ng/ml), total cholesterol 203 (N: <190 mg/dl) and low-density lipoprotein 136 (N: <100 mg/dl).

The results of red blood cell count, platelets, hemoglobin, thyroid function, blood coagulation were normal. Electrocardiogram showed sinus rhythm, 80 beats/minute, without signs of ischaemia. Her blood pressure was 130/85 mm Hg.

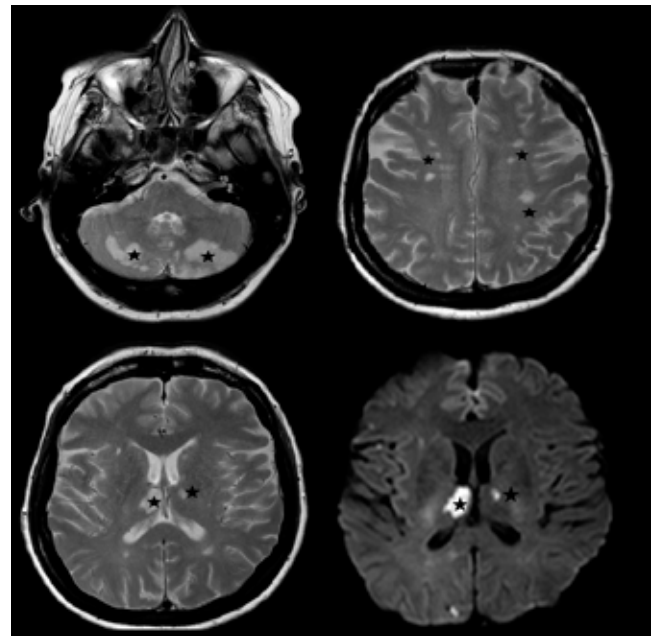


Figure 1. Cardiac magnetic resonance. Day 4. Upper-left: T2-weighted axial image with cerebellar stroke. Upper-right: T2-weighted axial image with diffuse ischemic changes in cerebral lobes. Lower-left: T2-weighted axial image with thalamic stroke. Upper-right: diffusion-weighted axial image with thalamic stroke

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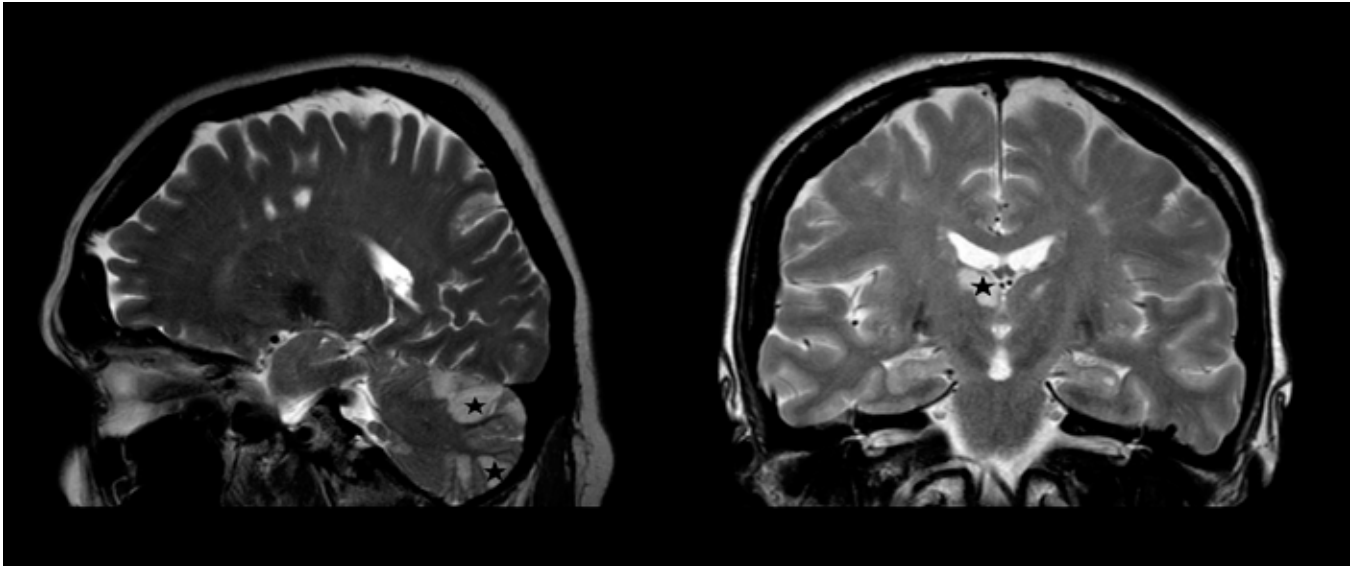


Figure 2. Cardiac magnetic resonance. Day 4. Left: T2-weighted sagittal image with cerebellar stroke. Right: T2-weighted coronal image with thalamic stroke



Figure 3. Transthoracic echocardiography. Left atrial myxoma (star). Left: Parasternal long axis view, diastole. Right: Apical four-chamber view, systole. Ao – aorta, LA – left atrium, LV – left ventricle, RA – right atrium, RV – right ventricle, MV – mitral valve

Computer tomography of the brain (without contrast) revealed multiple hypodense focal points in both cerebellar hemispheres, but more prominent in the left side. The image suggested microembolization, however, given the previous history of breast cancer we could not exclude the possibility of metastatic tumours. Magnetic resonance imaging showed multiple irregular areas in cerebellar hemispheres with increased signal in both T2-weighted imaging and diffusion weighted imaging. Similar areas were found in the cerebrum, thalamus, both frontal lobes, right temporal, right occipital and left parietal lobes (Figure 1, 2). These changes were not enhanced after contrast injection, which is typical for metastases. Bilateral scattered angio-derived changes were also observed.

The patient was diagnosed with multiple acute and subacute ischaemic changes in both cerebral and cerebellar hemispheres.

The multifocal character of the scattered changes suggested an embolic aetiology.

Doppler ultrasonography of carotid and vertebral arteries were unremarkable.

Echocardiographic examination was performed and revealed an irregular, multivillous, movable tumour of about 38×24 mm in size, localized in the left atrium, attached to the interatrial septum, suggesting the presence of a left atrial myxoma (Figure 3). Left ventricular contractility was normal with ejection fraction of 60%, mild mitral and tricuspid insufficiency and normal flow gradients were observed.

The patient was admitted to the department of cardiosurgery for further treatment. As a preoperative evaluation, coronarography was performed and excluded significant arteriosclerotic lesions

in the coronary arteries. The patient underwent surgical intervention and the tumour was successfully excised. The histopathologic examination confirmed the diagnosis of myxoma. Immediately following surgical treatment, the patient underwent cardiac rehabilitation and during a 6-month follow-up remains stable, with no symptoms of cardiac disease. Echocardiographic monitoring revealed no signs of recurrence.

Discussion

Primary cardiac tumours are relatively rare, they constitute merely 0,02% of all tumours found in autopsy [1], and 75% of them are benign [2]. Cardiac myxomas represent nearly 50% of all benign primary tumours of the heart found in autopsy. In the general population of patients undergoing a surgical excision of the cardiac tumour, 90% are benign with a dominance of myxomas (80%) [3]. The mean age of patients with myxoma is 50, however, myxomas may appear in any age group. Two thirds of patients are females. Myxomas in 75% of cases are localised in the left atrium, but may also arise in the right atrium (15–20%), ventricles, superior or inferior vena cava, and rarely, in pulmonary veins. The majority of myxomas originate in the atrial septum usually from the border of the fossa ovalis [4]. Multiple cardiac myxomas constitute about 5–7% of all cases [3,5] and are usually a part of familial autosomal dominant syndromes (Carney's complex, tuberous sclerosis, Gorlin's syndrome).

In macroscopic appearance, myxomas are polypoid structures, usually pedunculated, and rarely round or oval. The surface of the tumour is smooth or gently lobulated. Polypoid, smooth myxomas have little tendency to fragmentation. Uncommon papillary or multivillous myxomas tend to be fragile and may embolize frequently.

The differential diagnosis of cardiac tumour always includes the presence of thrombus, vegetation, and foreign body [4,5,12]. Myxomas may remain clinically silent, however, the clinical presentation varies and depends on the localization, size, and mobility of the tumour. Cardiac complications are usually related to mitral or tricuspid valve obstruction and present with symptoms of heart failure, dyspnoea, orthopnoea, recurrent pulmonary oedema, arrhythmia, sporadic syncope, and sudden death. Furthermore, a mobile tumour moving between the atrium and the ventricle may destroy the atrioventricular valve or subvalvular apparatus, causing severe mitral or tricuspid insufficiency.

Other unspecific systemic symptoms include fatigue, fever, muscle weakness, myalgia, arthralgia, weight loss, incidental Raynaud's phenomenon, cyanosis or finger clubbing. The laboratory tests generally reveal anaemia, elevation in the erythrocyte sedimentation rate and serum C-reactive protein, hypergammaglobulinaemia, leukocytosis, thrombocytopenia or thrombocytosis. Frequently, the first and only presentation of the disease, affecting 30 to 40% of patients with cardiac myxomas may be a systemic or cerebral infarct, as was detected in our patient [2,4]. Peripheral embolization may be caused by thrombus formation, but also by tumour fragments [6–9]. The diagnosis is confirmed by histologic examination after surgical removal of the embolus, which is rarely possible.

Early diagnosis of cardiac myxomas may allow for prevention of recurrent embolization. Transthoracic echocardiography is the diagnostic procedure of choice. If the result remains ambiguous, transesophageal echocardiography, computed tomography or nuclear magnetic resonance imaging may provide additional information. Surgery is the method of choice in the treatment of myxomas for the majority of patients and should be performed immediately after diagnosis to reduce thromboembolic risk. To avoid recurrence, a total full thickness excision of the tumour with adjacent interatrial septum is performed [11] and any arising septal defect is closed by direct suture or supplied with a patch made of pericardium or Dacron fabric. The risk of recurrence is 1 to 3% in sporadic tumours and 12 to 22% in familial or complex myxomas [2,4,10]. An echocardiographic examination performed every 6 months after surgery is recommended.

Secondary (metastatic) tumours of the heart are 20 to 40 times more frequent than primary tumours [4,12]. Moreover, a patient with a history of cancer and diagnosis of heart tumour naturally is suspected of having metastases to the heart. Fortunately, in our patient the tumour was an operable benign myxoma.

References

1. Reynen K. Frequency of primary tumors of the heart. *Am J Cardiol* 1996;77:107.
2. Braunwald Choroby Serca. Zipes, Libby, Bonow, Braunwald, red. wyd. pol. Banasiak W, Opolski G, Poloński L. Elsevier Urban & Partner Wrocław 2007, wyd. 1.
3. Bruce CJ. Cardiac tumors: diagnosis and management. *Heart* 2011;97:151–160.
4. Reynen K. Cardiac myxomas. *N Engl J Med* 1995;333:1610–1617.
5. Lee WC, Huang MP, Fu M. Multiple intracardiac masses myxoma, thrombus or metastasis: a case report. *J Med Case Rep*. 2015;9:179.
6. Lee VH, Conolly HM, Brown RD Jr. Central nervous system manifestations of cardiac myxoma. *Arch Neurol* 2007;64:1115–1120.
7. Sha D, Fan G, Zhang J. Multiple cerebral infarction as the initial manifestation of left atrial myxoma: a case report and literature review. *Acta Cardiol* 2014;69:189–192.
8. Kierdaszuk B, Gogol P, Kolasa A. Multiple metastatic intracranial lesions associated with left atrial myxoma. *Pol J Radiol*. 2014;79:262–267.
9. Kohno N, Kawakami Y, Hamada C. Cerebral embolism associated with left atrial myxoma that was treated with thrombolytic therapy. *Case Rep Neurol* 2012;4:38–42.
10. McCarthy PM, Piehler JM, Schaff HV, et al. The significance of multiple, recurrent, and "complex" cardiac myxomas. *J Thorac Cardiovasc Surg* 1989;91:389–396.
11. Waller DA, Ettles DF, Saunders NR, Williams G. Recurrent cardiac myxoma: the surgical implications of two distinct groups of patients. *Thorac Cardiovasc Surg* 1989;37: 226–230.
12. Reynen K, Köckeritz U, Strasser RH. Metastases to the heart. *Ann Oncol* 2004;15:375–381.